Multiple Dural Arteriovenous Fistulas
Radiologic Progression and Endovascular Cure
Case Report

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Summary

Dural arteriovenous fistulas are most probably acquired lesions. However, they have been rarely encountered de novo. We present a unique case of a 71-year-old woman who initially presented with right-sided dural arteriovenous fistula (DAVF), which spontaneously resolved after diagnostic arteriography. She later developed asymptomatic occlusion of the left transverse sinus. Five years after her initial presentation she developed left-sided pulse-synchronous tinnitus. MRA and catheter angiography showed a complex type IV DAVF between the left transverse sinus and multiple dural branches arising from both left and right external carotid arteries. The left transverse sinus was isolated from the torcula herophili, with stenosis of the sigmoid sinus. Extensive cortical venous drainage was demonstrated. Endovascular cure was effected by polyvinyl alcohol particle and absolute alcohol occlusion of the dominant dural supply, and transvenous coil occlusion of the left transverse sinus. The patient's symptoms resolved almost immediately.

This unique case demonstrates that dural sinus occlusion and DAVFs may co-exist, but there may not be a causal relationship. It is likely that both DAVFs and sinus occlusion are manifestations of the same disease process characterised by a pro-thrombotic state and secondary angiogenesis. It is important to recognise that changes in symptomatology, even long after apparent disappearance of a lesion may indicate recurrence, and careful follow up is advocated.

Introduction

Dural arteriovenous fistulas are considered a venous disease. However, the relationship between dural shunts and pathology of the dural sinuses is complex. Several case reports have documented the appearance of DAVFs as evidence that they are acquired lesions, typically occurring in middle age in relation to occlusion (thrombotic, surgical or tumoral) of one or more of the major dural venous sinuses [1-14].

Progression of DAVFs from low-risk to high-risk lesions with cortical venous drainage has been reported in longitudinal studies [15-18]. However, the spontaneous appearance and resolution of a low grade DAVF, with subsequent appearance at another site of a more dangerous lesion five years later associated with sinus occlusion has not been previously reported. Such a sequence of events is indicative of an acquired lesion with a tendency towards progression, even over several years.

The importance of careful patient follow-up with particular reference to recurrence of symptoms cannot be overemphasised, even in low-risk lesions considered to have resolved.

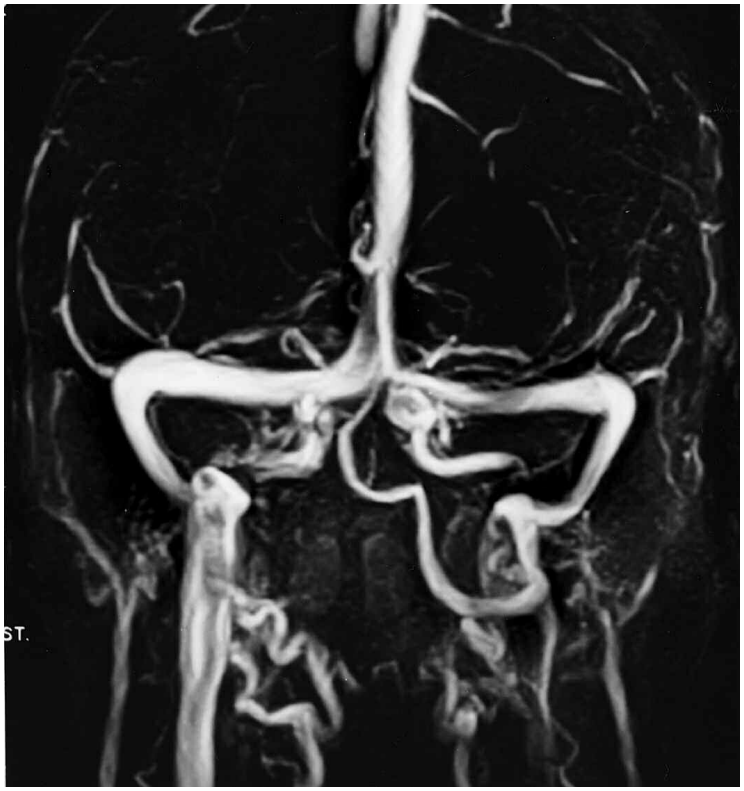


Figure 1 2D time of flight magnetic resonance angiography at initial presentation demonstrates abnormal proliferation of vessels around the right jugular bulb. Note patency of all the major venous sinuses.

Case History

A previously fit 66-year-old lady developed pulse-synchronous tinnitus in august 1995. This was audible with a stethoscope and prominent middle ear vessels were noted on otoscopy. MRI, MRA and MRV were performed in october 1995 revealing abnormal vessels in relation to an enlarged right jugular bulb (figure 1). In view of the symptoms and the clinical suspicion of a DAVF, cerebral angiography was performed in march 1996. A grade I DAVF between the right occipital artery and the ipsilateral jugular bulb was demonstrated with rapid arteriovenous shunting (figure 2). There were no adverse features to suggest that this was a high-risk lesion and the major venous sinuses were patent.

A few hours after the angiogram, the patient became hypotensive, nauseated and vomited. These symptoms spontaneously resolved and she subsequently noticed abrupt cessation of her pulsatile tinnitus. Thereafter she remained well and was discharged from hospital. The symptoms were clearly vasovagal and possibly related to a sudden vascular event: either occlusion of the right-sided DAVF (supported by cessation of symptoms) or occlusion of the left transverse sinus. However, follow-up imaging was not performed until eight months later.

2D Time of Flight (TOF) MR angiography was performed in november 1996, while the patient remained asymptomatic (figure 3). This showed no abnormal vessels in relation to the right jugular bulb. However, the left transverse sinus was not visible and was interpreted as being completely occluded.

In February 2001, the patient re-presented with left-sided pulse-synchronous tinnitus. MRI, MRA and MRV performed urgently demonstrated abnormal vessels over the left cerebral convexity and in relation to the left skull base (figure 4).

The left transverse sinus remained occluded proximally, but there was a patent segment laterally. The findings were confirmed at angiography to be due to a complex type IV dural arteriovenous fistula. Multiple dural arteriovenous shunts involving three segments of a partially occluded and irregular left transverse sinus were identified: proximally from the right occipital artery (figure 5A); distally from the

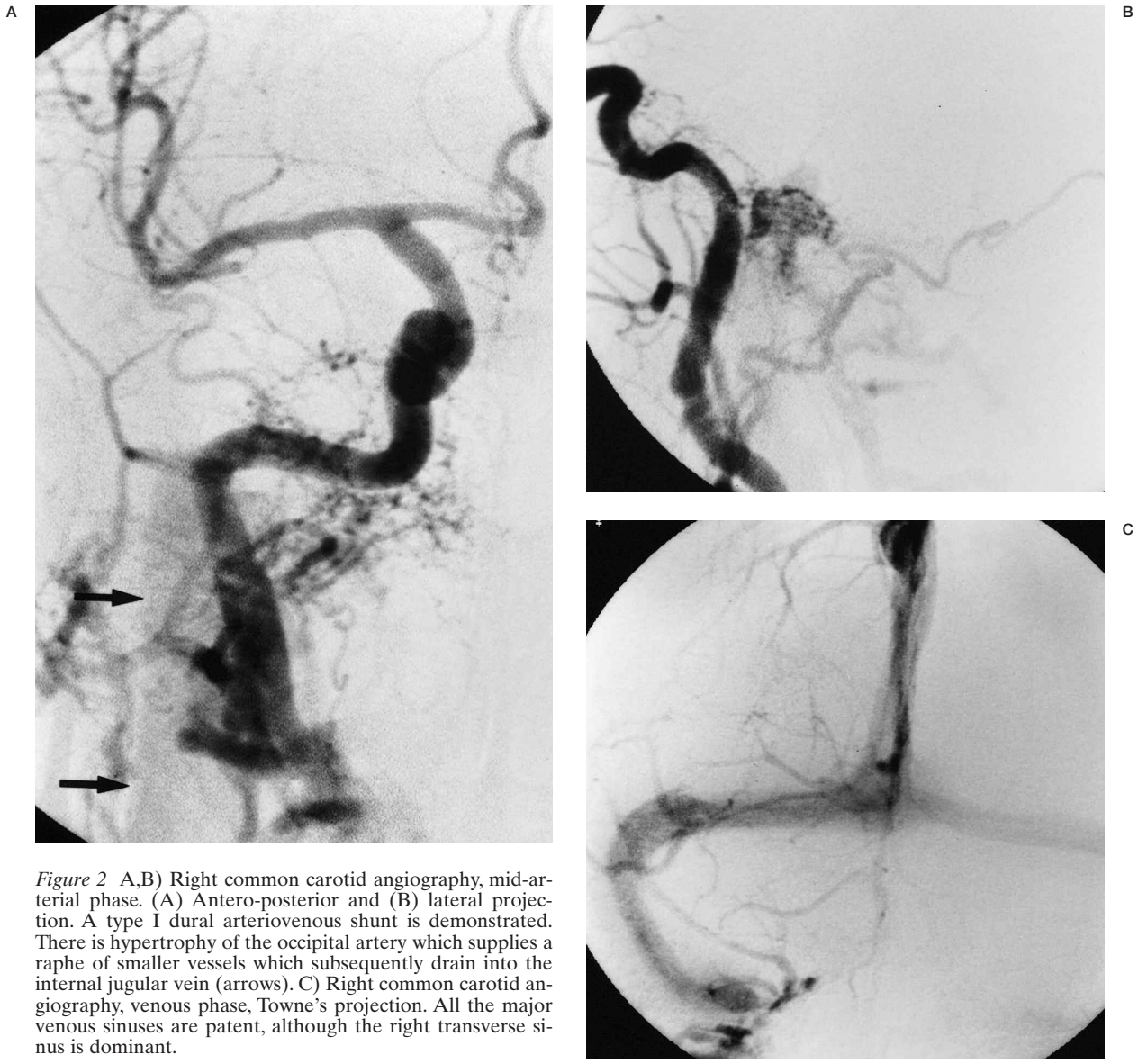


Figure 2 A,B) Right common carotid angiography, mid-arterial phase. (A) Antero-posterior and (B) lateral projection. A type I dural arteriovenous shunt is demonstrated. There is hypertrophy of the occipital artery which supplies a raphe of smaller vessels which subsequently drain into the internal jugular vein (arrows). C) Right common carotid angiography, venous phase, Towne's projection. All the major venous sinuses are patent, although the right transverse sinus is dominant.

left occipital artery, left middle meningeal artery and left posterior auricular artery (figure 5B); and into the left sigmoid sinus from multiple vertebrobasilar meningeal branches via a parallel venous channel (figure 5C). Extensive cortical venous reflux was demonstrated in relation to the shunt at the lateral aspect of the left transverse sinus. The left sigmoid-jugular junction was stenosed. There was no filling of the left transverse sinus on carotid angiography.

The angiographic features were compatible with partial recanalisation of the left transverse sinus, which was anatomically isolated from the torcula herophili.

There was abnormal arteriovenous delay and beading of cortical venules as evidence of venous hypertension. Extensive anterior and deep venous drainage was shown to the cavernous and straight sinuses respectively.

The DAVF had clearly progressed to a dangerous lesion and endovascular obliteration was undertaken in view of the risk of intracranial hemorrhage and venous ischaemia, rather than her symptoms, which were not disabling.

Initial angiographic assessment during the

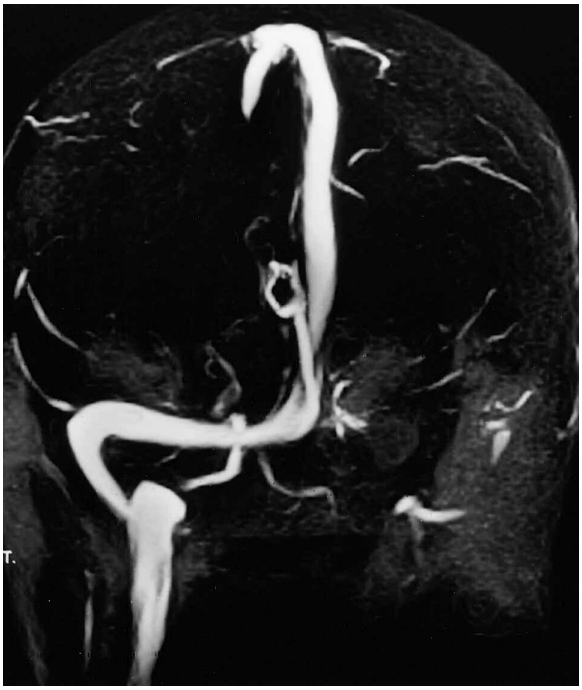


Figure 3 MR venogram performed 8 months after resolution of symptoms reveals occlusion of the left transverse sinus. There is no pathological vascular proliferation.

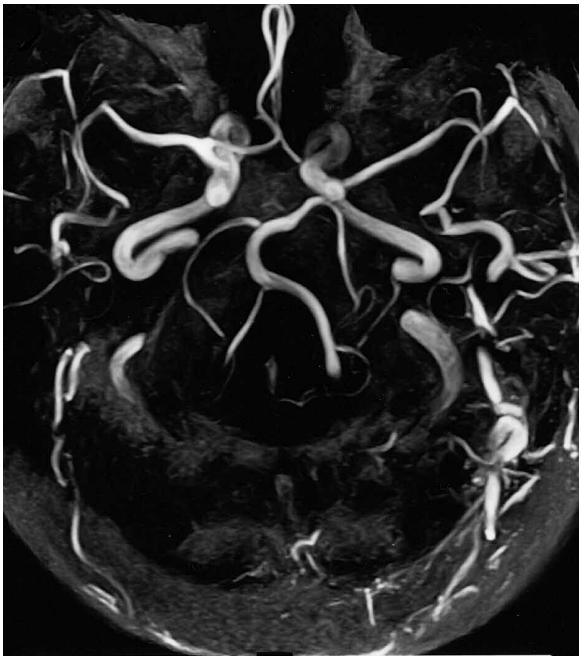


Figure 4 2D time of flight magnetic resonance angiography at re-presentation in february 2001 demonstrates proliferation of vessels around the left transverse-sigmoid junction. MRV demonstrated occlusion of the left transverse sinus. Axial source images confirmed patency of the mid left transverse sinus.

embolisation procedure two months later showed an increase in the degree of arteriovenous shunting and cortical venous reflux. Additionally, a venous pouch was visible arising from the distal aspect of the left transverse sinus draining retrogradely into multiple cortical veins (figure 6).

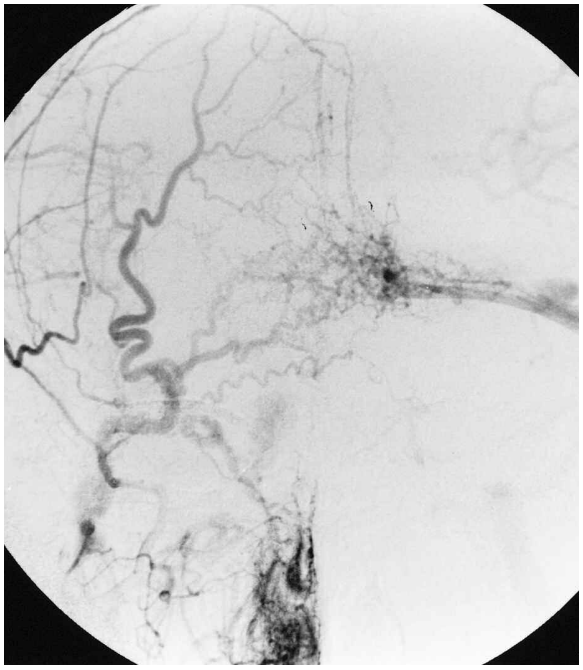
The right and left occipital arteries and the left posterior auricular artery were embolised with polyvinyl alcohol (PVA) particles 250-355 micron size. Absolute alcohol was also injected into the left occipital artery (figure 7).

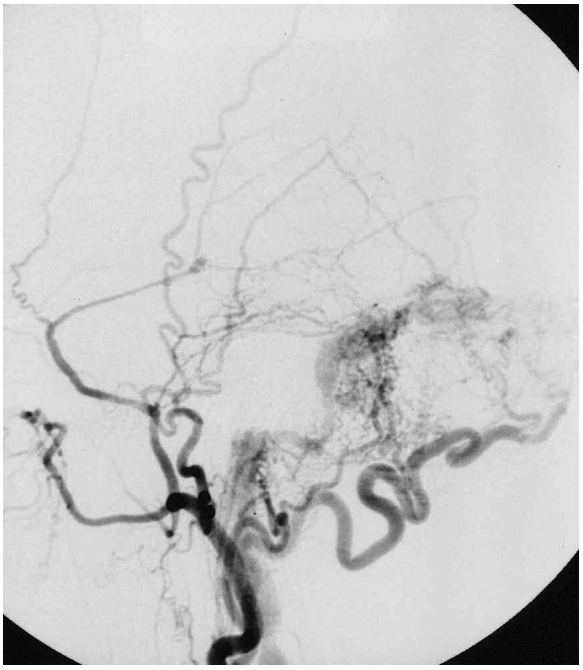
The left jugular vein was catheterised via a femoral approach. A guiding catheter was navigated with difficulty into the distal left transverse sinus through the stenosed segment. Embolization of the left transverse sinus was performed with fifty fibre coils sequentially placed from the point of fistulation of the right occipital artery medially, to the stenosis at the sigmoid-jugular junction. The venous pouch connecting with the cortical veins was also packed with coils. Final angiography (figure 8) demonstrated no remaining fistulation, with drainage of the entire intracranial compartment into the superior sagittal sinus and right transverse sinus, and anteriorly to the cavernous sinus and inferior petrosal sinus.

The patient remained neurologically intact with complete resolution of her pulsatile tinnitus. She suffered serous otitis media as a result of the prolonged general anaesthetic, which resolved spontaneously. At follow-up there was no evidence of cognitive dysfunction.

Discussion

The complex series of events in our patient may be explained on the basis of what is currently understood about these lesions from case series and pathological studies in the literature. There is evidence that spontaneous dural arteriovenous fistulas are acquired lesions that are often encountered in association with occlusion of one or more of the major venous sinuses. However, the role of sinus thrombosis and occlusion in the aetiology of DAVFs is complex. DAVFs have been shown to occur after sinus occlusion [4,8,9,11-14] and at the same time as sinus occlusion [27]. Additionally, sinus occlusion may be associated with progression of DAVFs [4,18] associated with spontaneous obliteration of DAVFs [29], and sinus occlusion [7] and re-





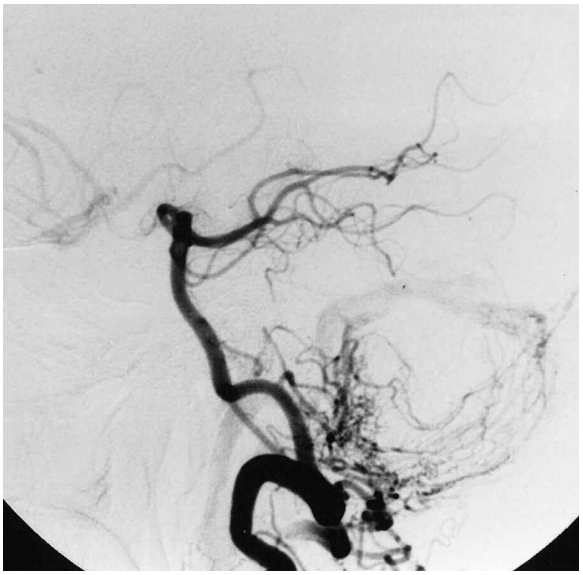


Figure 5 A) Right external carotid arteriography, late arterial phase, Towne's projection. There are shunts between the branches of the right occipital artery and the medial left transverse sinus. Cortical venous drainage is demonstrated due to restriction of exocranial venous outflow caused by the narrowed sinus. B) Left external carotid arteriogram, late arterial phase, lateral projection. There is shunting between branches of the occipital, middle meningeal and posterior auricular artery to the mid left transverse sinus. A small flow-related aneurysm is demonstrated in relation to the anterior division of the middle meningeal artery. C) Left vertebral angiogram, early arterial phase, lateral projection. Multiple meningeal branches from the vertebrobasilar system show shunting into a small parallel venous channel, which drains into the distal left transverse sinus.

canalisation [25,28] have been used to treat DAVFs.

The initial lesion presented without evidence of sinus occlusion or stenosis and there was no evidence angiographically of prior sinus compromise. Dural arteriovenous shunts are not always associated with dural sinus compromise [19] and DAVFs are not inevitable after sinus occlusion [9]. However, in the case reported by Kawaguchi [19], the DAVF occurred after embolisation of a carotico-cavernous fistula at a different site; evidence for a more generalised venous disorder which predisposes the patient to multiple dural arteriovenous shunts.

The clinical presentation of pulse synchronous tinnitus is typical for lesions in the region of the jugular bulb [20-23]. However, the temporal relationship of the recovery of the patient's symptoms (and presumed occlusion of the DAVF) to occlusion of the left transverse sinus is uncertain, as spontaneous closure of a DAVF may occur without sinus occlusion [30]. The pathological study by Nishijima et Al [24] may provide the explanation; spontaneous closure of a fistula may occur due to the progressive thickening of the intima of mural arteries and veins to such a degree that the lumina are occluded. The role of diagnostic angiography in the cessation of symptoms is unclear. However, the possibility of microemboli obliterating a critical number of feeding arterioles, or thrombosis induced

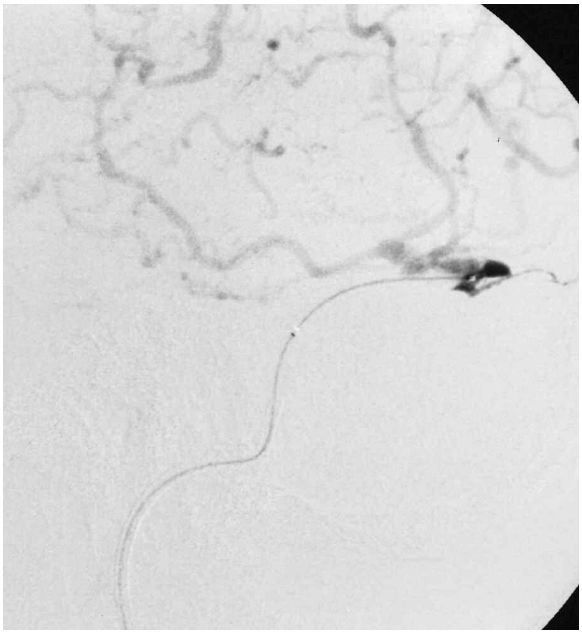


Figure 6 Retrograde transvenous approach to the left transverse sinus allowed passage of a microcatheter into the venous pouch demonstrated in fig 5A. Extensive cortical venous drainage is demonstrated.

by contrast medium injection are speculative possibilities.

Occlusion and partial recanalisation of the left transverse sinus preceded the development of the left-sided DAVF by up to 5 years. Occlusion of the left transverse sinus appeared to be clinically silent. Such an apparent lead-time is well recognised. Sakaki et Al demonstrated an interval of 22 to 73 months from sinus occlusion and/or excision to the development of a new DAVF [8]. However, in their series, DAVFs occurred in only 5 of 69 (7%) of patients where the sigmoid and transverse sinuses had been operated on; evidence for a necessary additional trigger. The effect of sinus occlusion on venous pressure may be an important factor in the genesis of DAVFs [26]. Additionally, the role of chemical mediators such as basic fibroblast growth factor (bFGF) have been have been studied. Terada et Al found strong staining for bFGF within specimens from patients with DAVF and no staining with a control specimen [31]. A clear-cut causal link is currently theoretical. An underlying biochemical de-

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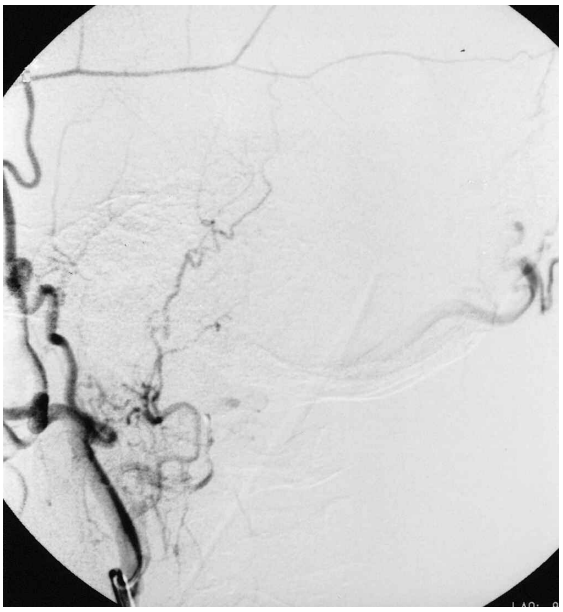

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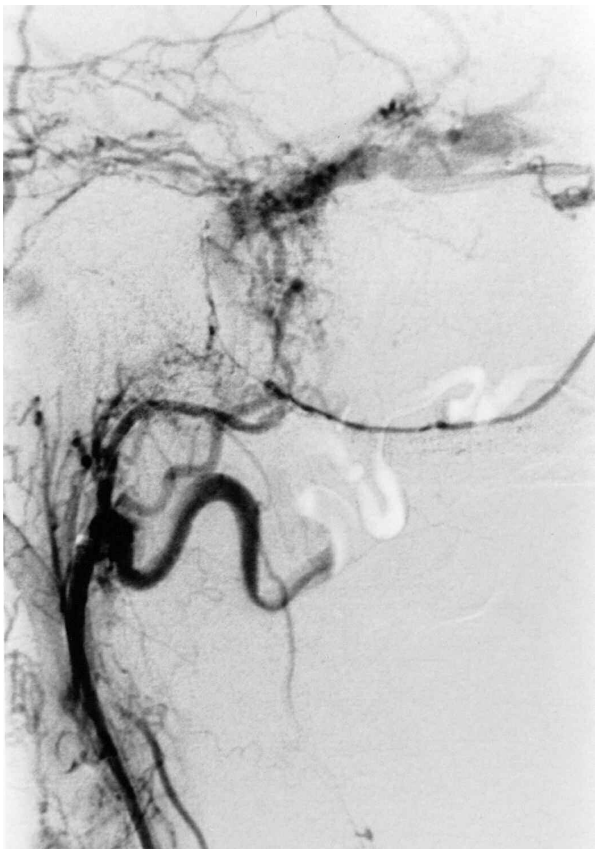

Figure 7 A) Right and (B) left external carotid arteriogram after polyvinyl alcohol (PVA) and absolute alcohol injection of the occipital arteries.

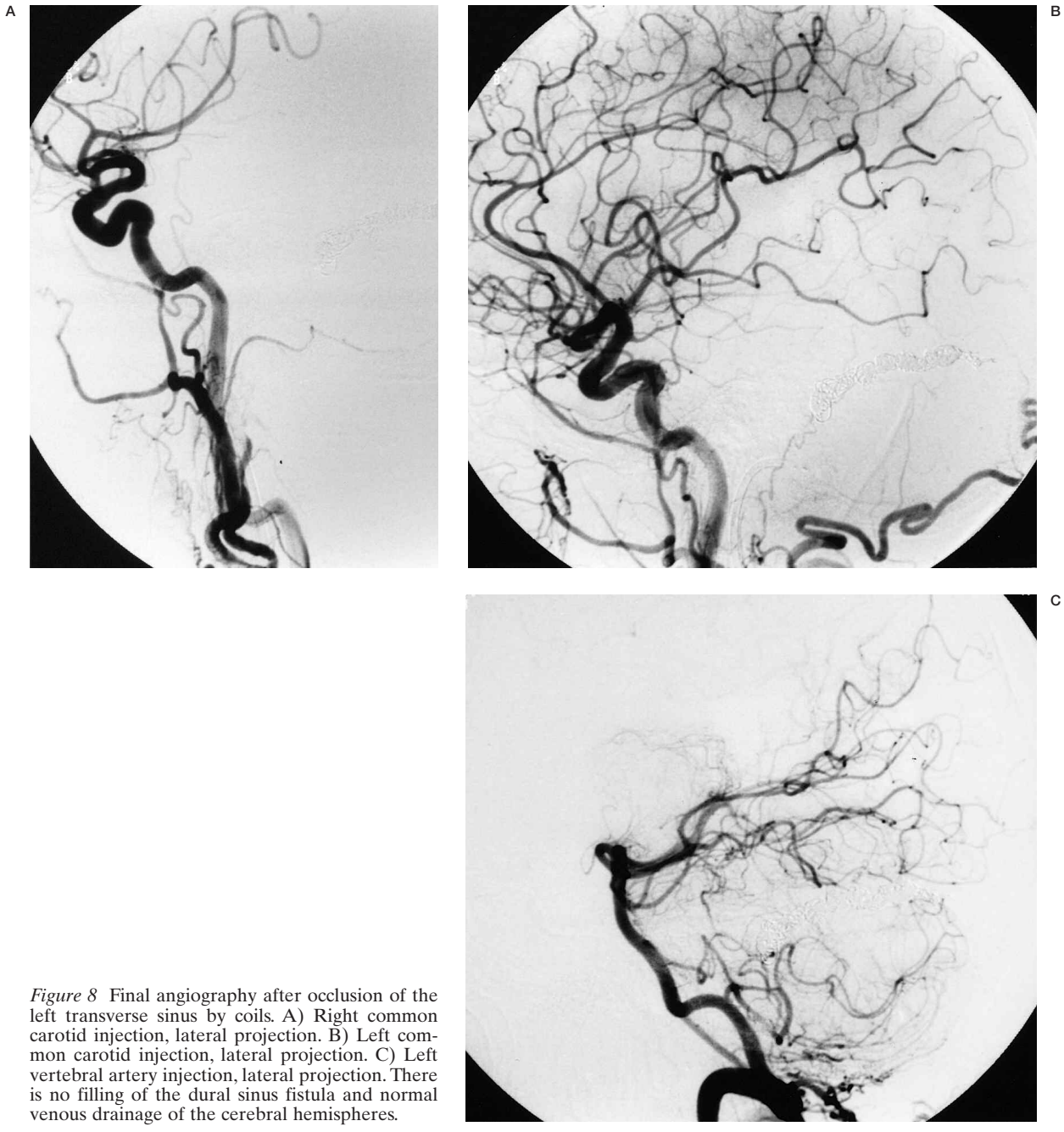

Figure 8 Final angiography after occlusion of the left transverse sinus by coils. A) Right common carotid injection, lateral projection. B) Left common carotid injection, lateral projection. C) Left vertebral artery injection, lateral projection. There is no filling of the dural sinus fistula and normal venous drainage of the cerebral hemispheres.

rangement may provide an explanation for the development and progression of the DAVF in our patient.

The complex interrelationship between DAVFs and sinus disease has been explored by Nishijima et Al [24], who analysed the histology of normal sinuses and those involved in DAVFs. Numerous arterioles and venules are present within the normal sinus wall, the venules occasionally communicating with the sinus lumen, but without direct arteriovenous connections. Patients with DAVFs showed evidence of sinus thrombosis and recanalisation with thickening of the intima and adventitia of the dural arteries with luminal stenosis and occlusion. Dural veins were dilated and communicated freely

with one another and the dural venous sinus itself. Additionally, fistulous connections between the dural arteries and veins, but not directly between the arteries and the venous sinuses were demonstrated. Dural arteriovenous shunting therefore may be an indirect consequence of sinus thrombosis. This appears to be the sequence of events in our case. However, the lumen of the dural sinuses may be further narrowed by a combination of direct impingement by the arteriovenous anastomoses within with dural sinus wall, severe sinus intimal thickening and partial thrombosis and recanalisation [24]. These pathologic findings have been confirmed by other studies, although sinus thrombosis has been shown only inconsistently [8]. Thus a cycle of anatomic and pathologic changes occurs with the potential for disease progression over time. Additionally, venous hypertension appears not only to be a consequence of dural shunts, but is a necessary component in the generation of AV shunts. Animal studies have shown that venous hypertension in addition to sinus occlusion are required for the formation of DAVFs [26]. Once venous hypertension develops, then the self-perpetuating cycle of events leading to disease progression is induced.

The co-existence of sinus occlusion and arteriovenous shunts in the sinus wall necessitates diversion of flow to leptomeningeal veins creating venous hypertension and increasing the risk of intracranial hemorrhage. Our case demonstrated an isolated segment of dural sinus with direct fistulation between dural arteries, the sinus lumen and cortical veins, which supports the pathological findings described. Sinus occlusion associated with DAVF has a significant negative correlation with spontaneous resolution [16]. The anatomy of the DAVF and the clinical expression of the disease may be in part determined by the efficiency of sinus recanalisation and the anatomical relationship of the dural shunts to occluded and patent segments of the venous sinuses.

Our patient's subsequent angiographic and clinical progression is unusual and suggests that even if a low-grade lesion spontaneously resolves, the anatomy and physiology of the venous sinuses may be permanently altered, with increased susceptibility to the development of further lesions.

Conclusion

The varied nature of the angioarchitecture, clinical expression and prognosis of DAVFs may be explained by a number of complex processes that occur during the course of the disease. The anatomical basis for the development of DAVFs is the presence of arterioles and venules within the wall of the sinus, but the trigger is as yet unknown. As the arteriovenous fistulas develop, the vessels become thickened and may thrombose resulting in spontaneous closure of the fistula. Alternatively, the fistulas may progress and result in stenosis of the sinus due to a combination of vessel engorgement and mural intimal thickening. The result may be occlusion of the sinus adjacent to the lesion. This may subsequently result in occlusion of the DAVF. Alternatively, if the mural venules communicate with cortical veins, the fistula may not close and be redirected to the cortical veins resulting in a dangerous progression of the lesion. Thus, the outcome of patients with DAVFs is difficult to predict and depends on individual lesion angioarchitecture. If is therefore imperative that patients are followed up closely and any change in symptoms should alert the clinician to a potential deterioration or recurrence of the lesion, even after several years.

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